

Outcomes in patients with interrupted aortic arch and associated anomalies: a 20-year experience

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Abstract

Objective: The surgical results for the repair of interrupted aortic arch (IAA) have evolved in recent years. We report our results for staged repair of this complex congenital malformation. **Methods:** Sixty-five patients (mean age, 16.9 ± 41.7 days) were diagnosed with IAA and referred for surgical therapy. The surgical management strategy at our institution between 1982 and 2005 has been one-stage complete repair ($n = 13$) or staged repair ($n = 52$) in selected patients. Non-complex patients (group I, $n = 51$) had a ventricular septal defect (87%), aortopulmonary window (8%), and left ventricular outflow tract obstruction (27%). Group II ($n = 14$) were patients with Taussig–Bing double outlet right ventricle ($n = 6$) or truncus arteriosus ($n = 8$). Method of staged repair of IAA was to transect and turn down the left carotid artery and anastomosis it to the descending aorta ($n = 41$) or graft interposition ($n = 2$) combined with a pulmonary artery (PA) banding followed in a few months by delayed ventricular septal defect (VSD) closure and PA de-banding. **Results:** There were 5 early and 10 late deaths. The actuarial survival including early mortality was 92% at 1 year, 81% at 5 years, and 76% at 10 and 15 years. There was an 81% 15-year survival for children in group I compared with a 54% for children in group II ($p < 0.001$). Risk factors for increased mortality by univariate analysis were as follows: (1) primary aortic anastomosis ($p = 0.03$), (2) presence of complex anomalies ($p = 0.05$), and (3) initial IAA repair performed before 1994 ($p = 0.05$). Actuarial freedom from any type of aortic reoperation or intervention was 86% at 1 year, 69% at 5 years, and 60% at 10 and 15 years. Univariate and multivariate analyses identified no tested variables as risk factors for reoperation. The majority (86%) was in New York Heart Association (NYHA) class I, and 14% remained in NYHA class II. During the postoperative course there were no neurologic deficits, seizures, and growth disturbances in any patient. **Conclusion:** Staged repair of IAA using a left carotid artery turn down can be safely applied in IAA patients with and without other intracardiac anomalies with good results. Use of the left carotid artery for arch reconstruction did not result in any detectable neurological events or growth disturbances later in life. Associated anomalies played an important role in outcomes. The long-term probability for reoperation and/or reintervention remains high regardless of operative technique.

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1. Introduction

Interrupted aortic arch (IAA) is an uncommon congenital cardiovascular malformation characterized by the lack of continuity between the ascending and descending thoracic aorta as described by Steidele in 1778 [1]. If IAA is untreated, the median age at death is 4–10 days, usually following physiological closure of the ductus arteriosus [2].

IAA may be associated with a variety of other more complex cardiovascular anomalies, such as truncus arteriosus, aortopulmonary window, double-outlet right ventricle, and transposition of the great arteries. Obstruction or

hypoplasia of the left ventricular outflow tract (LVOT) is also common. DiGeorge syndrome occurs in more than 25% of these patients.

During the last several years, results of both the one-stage and two-stage repairs of IAA have improved, and the early mortality in some centers approached 10% [3–5]. The long-term outcomes of survivors after one-stage or staged approaches have not been extensively reported. A high rate of development of restenosis at the site of aortic anastomosis, left ventricular outflow tract obstruction (LVOTO) and left bronchial compression have been reported [3,4,6,7]. Other residual lesions and non-cardiac complications have been described that often require reoperation and represent a risk of late mortality [2,5,8–10].

The long-term results comparing complete repair versus staged correction has not been reported. Our philosophy has

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advocated staged repair of IAA when possible, especially in the presence of complex-associated cardiac anomalies. In this report, we review our 20-year experience comparing staged with primary total correction of interrupted aortic arch.

2. Materials and methods

2.1. Patients

Between November 1982 and July 2005, 65 consecutive neonates with diagnosis of IAA and ventricular anatomy amenable to biventricular repair underwent operation at the James Whitcomb Riley Hospital for Children in Indianapolis, IN, USA. We reviewed the medical records with regard to the initial clinical features, pathophysiological findings, surgical treatment, and hospital mortality after obtaining approval from the Indiana University Institutional Review Board. Data from outpatient visits and from patients dying after hospital dismissal were obtained from attending physicians, hospitalization records, or death certificates.

There were 39 boys and 26 girls. Their mean age and body weight at the initial operation were 17.1 ± 40.4 days (range, 1–210 days) and 3.3 ± 0.5 kg (range, 1.5–4.5 kg). All patients were in congestive heart failure receiving prostaglandin infusion. Fifty-two percent were mechanically ventilated. Preoperative resuscitation, necessary in 46 children, included prostaglandin E1, assisted ventilation, inotropic support and diuretics. DiGeorge syndrome was diagnosed in 17 patients (26%; 2 with IAA type A and 15 with IAA type B). Truncus arteriosus of types I, II, and III truncal anatomy was seen in five, one, and two children, respectively [11].

All patients underwent preoperative cardiac echocardiography or catheterization, or both. Cardiac catheterization was performed in 14 patients to confirm the echocardiographic findings and associated defects. The anatomic features of the aortic arch and the site of interruption were determined according to classification of Celoria and Patton [10]. There were 12 patients with type A, 51 with type B, and 2 with type C. A retro-esophageal left subclavian artery was present in 14 patients.

The intracardiac- and extracardiac-associated lesions are listed in Table 1. On the basis of the anatomic features, patients with IAA were divided into group I (non-complex): 51 patients with associated defects including patent ductus arteriosus (PDA), ventricular septal defect (VSD), aortopulmonary window, atrial septal defect (ASD), and various forms of LVOTO (valvar aortic stenosis, subaortic obstruction, supravalvar aortic stenosis); and group II (complex): 14 patients with truncus arteriosus ($n = 8$) or Taussig–Bing double-outlet right ventricle (DORV; $n = 6$).

2.2. Operative procedures

One-stage complete repair was performed in 13 patients (group I, 7 children; group II, 6 children) while 52 underwent staged reconstruction (group I, 44 children; group II, 8 children) (Fig. 1).

In 13 patients the one-stage IAA repair and associated heart lesions were performed from the midline sternotomy approach with extracorporeal circulation, aortic cross-

Table 1
Intracardiac- and extracardiac-associated lesions in patients with IAA

	Group I ($n = 51$)	Group II ($n = 14$)
PDA	50	13
VSD	44	13
ASD	22	10
AS	11	0
Subaortic AS	6	3
Multilevels of LVOTO	7	0
AP window	4	0
Truncus arteriosus	0	8
Taussig–Bing DORV	0	6
DiGeorge syndrome	15	2
Goldenhar syndrome	2	0

AP: aortopulmonary; AS: aortic stenosis; ASD: atrial septal defect; DORV: double-outlet right ventricle; IAA: interrupted aortic arch; LVOTO: left ventricular outflow tract obstruction; PDA: persistent ductus arteriosus; VSD: ventricular septal defect.

clamping, and cardioplegia. The repair consisted of direct end-to-end anastomosis between the descending and the ascending aorta ($n = 11$) or interposition of a polytetrafluoroethylene (PTFE) tube (W.L. Gore & Associates Inc., Flagstaff, AZ, USA) in two patients (with type C IAA), and correction of associated heart lesions: ligation of the PDA ($n = 11$), atrial and ventricular septal defects closure ($n = 8$), truncus arteriosus repair with the conduit ($n = 5$), arterial switch procedure ($n = 1$), and others ($n = 4$).

Staged reconstruction of the aortic arch was performed by anastomosing the divided end of the left carotid artery to the descending aorta, ligation of the patent ductus arteriosus and pulmonary artery banding employing a fourth inter space left thoracotomy. In all patients, blood pressure in the ascending aorta was monitored proximal to the site of the occluding clamp on the transverse aortic arch. Usually, the right radial artery was cannulated, but because the right subclavian artery originated from the descending aorta in 14 patients with type B interruption, right superficial temporal artery monitoring was also used when necessary. A blood pressure cuff was placed on the lower extremity.

The innominate artery, transverse aortic arch, left subclavian artery, and ductus arteriosus were circumferentially dissected. The descending thoracic aorta was completely mobilized for five to eight pairs of intercostal arteries. Special care was taken to mobilize the left common carotid artery as far cranially as possible. Vascular clamps were applied and the upper extremity blood pressure remained 60–80 mmHg. The ductus arteriosus was divided and after the removal of all ductus tissue, three techniques were employed to repair the interruption: (1) in 43 patients, the left common carotid artery was divided, spatulated appropriately and anastomosed to the descending thoracic aorta with absorbable suture (Fig. 2A and B), (2) direct anastomosis between the ascending and descending aortic segments was employed in seven patients (Fig. 3A and B), and (3) synthetic patch aortoplasty of a primary anastomosis in two patients was performed. Following aortic reconstruction, the pulmonary artery was banded to reduce the distal pulmonary artery pressure to 50% or less of the systemic value.

The second stage consisted of prosthetic patch closure of the VSD and pulmonary artery debanding with pulmonary

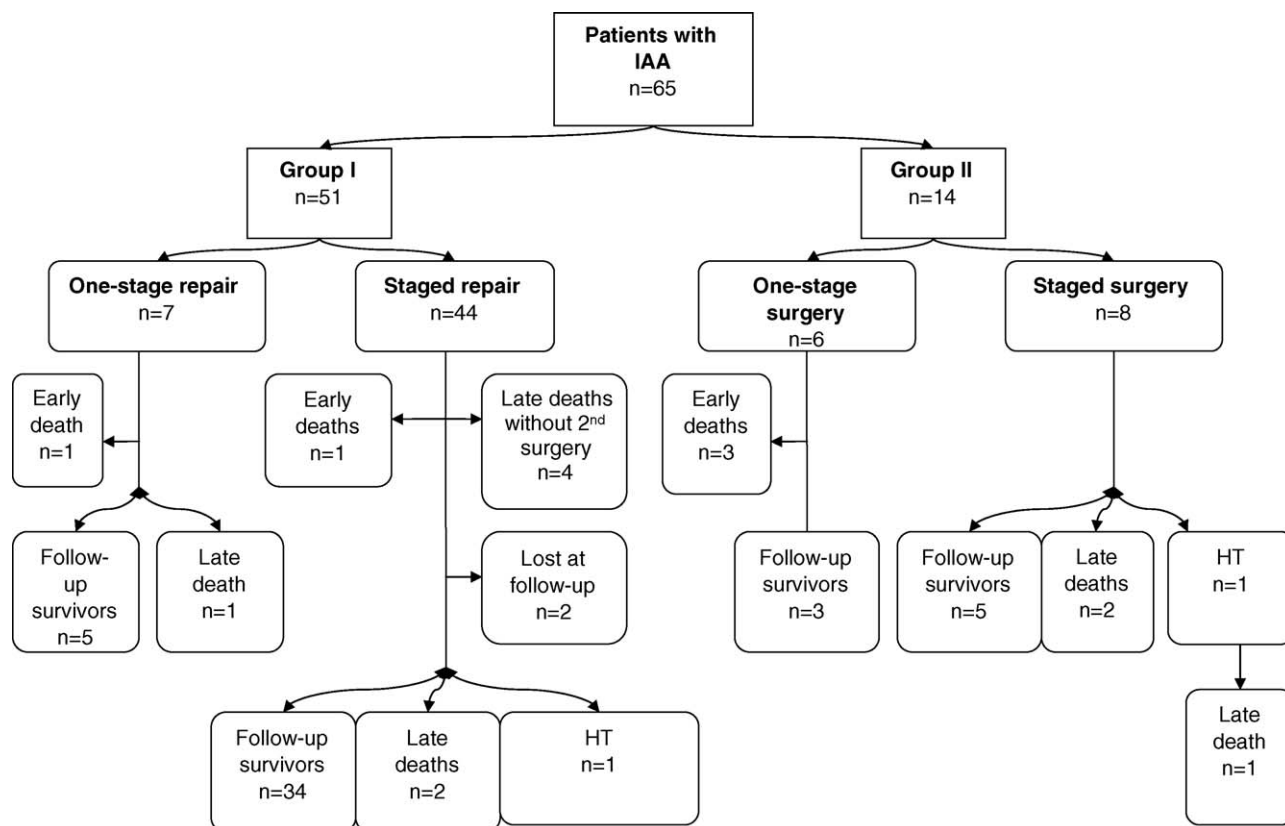


Fig. 1. Distribution and disposition of patients with interrupted aortic arch who underwent various surgical repairs (HT: heart transplantation).

artery reconstruction employing PTFE or autologous pericardial patch. The median age at the second operation was 10 months (range, 7 days to 7 years). Four of the 52 patients (8%) had the second stage performed within 2 weeks of the first stage because of persistent heart failure or pulmonary artery distortion by the PA band. In all patients, the mean cross-

clamp time was 88 ± 36 min (range, 53–176 min) and the mean bypass time was 143 ± 46 min (range, 93–251 min). The VSD was generally closed through a right atriotomy and occasionally by a small right ventriculotomy. Two patients required pacemaker insertion following VSD closure. Cardiopulmonary bypass was continued throughout the repair and was rarely interrupted during the intracardiac repair. Modified ultrafiltration was performed after the final stage of rewarming and discontinuation of bypass.

LVOTO was generally the result of a posterior misalignment of the septal conus and was assessed at echocardiography by measurement of the subaortic diameter, the aortic annulus diameter, and the ascending aortic diameter. When the subaortic diameter was smaller than two-thirds of the aortic annular diameter, subaortic stenosis was considered to be severe. If LVOTO obstruction was considered to be mild, the surgery for LVOTO relief was not performed or was delayed. Preoperative mean LVOTO gradient ($n = 27$) was 20.7 ± 9.7 mmHg (range, 8–44 mmHg). One patient in group I underwent closed transventricular aortic valvotomy. Four patients in group I with a preoperative diagnosis of severe LVOTO had resection of obvious obstructing subaortic tissue or valvotomy for valvar commissural fusion, or a combination during the second stage within 2 weeks after the initial IAA repair in the same hospital stay.

Arterial switch with VSD closure was performed in four patients with Taussig–Bing and DORV. A Lecompte maneuver was utilized in all patients. Two patients with subaortic obstruction in group II underwent Rastelli-type conduit

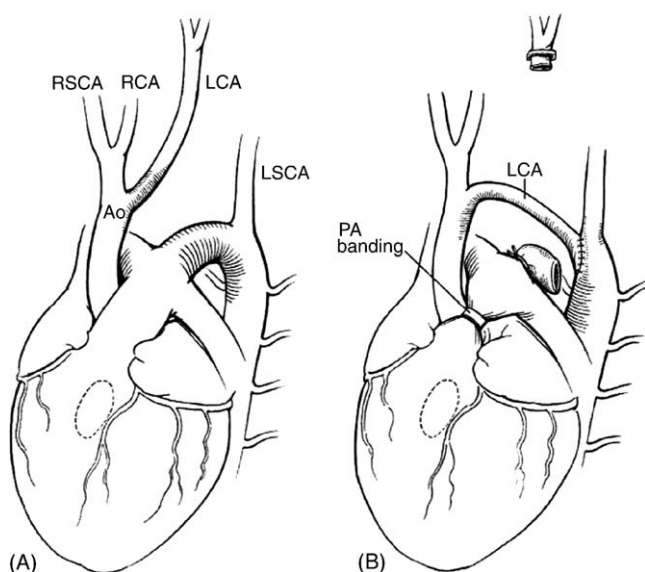


Fig. 2. Carotid artery swing-down repair of interrupted aortic arch; before (A) and after (B) correction.

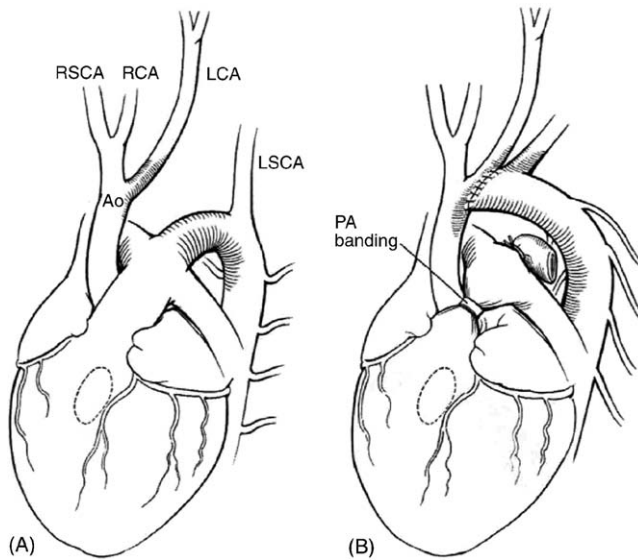


Fig. 3. Direct end-to-end anastomosis of interrupted aortic arch repair; before (A) and after (B) correction.

reconstruction and a Damus–Kay–Stansel procedure. Truncus arteriosus repair was achieved with the Rastelli-type operation. The mean conduit size in patients of group II was 13 ± 2 mm (range, 11–18 mm).

2.3. Statistical analysis

SPSS statistical program for Windows version 10 (SPSS Inc., Chicago, IL, USA) was used to perform data analysis. Data are expressed as mean \pm SD and range. The Kaplan–Meier product limit and Cox proportional hazards regression methods were used for actuarial survival and freedom from reoperation analysis. Multiple regression analysis was performed as conditional backward stepwise proportional

hazards regression. p -values of ≤ 0.05 were considered significant. Early mortality is defined as hospital death or death within 30 days of discharge. All other death is considered as late mortality.

3. Results

3.1. Mortality

There were two early deaths in group I (4%; 2/51). One patient died of low cardiac output following carotid artery interposition and PA banding for type B IAA with VSD. A second death occurred in a 2-kg baby with type B IAA, VSD, ASD, and critical aortic stenosis who underwent direct anastomotic repair of IAA, patch closure of VSD and ASD, and closed transventricular aortic valvotomy. This patient expired secondary to acute renal failure 1.5 months postoperatively.

In group II, there were three early deaths (21%; 3/14). Two patients died of low cardiac output following repair of truncus arteriosus and direct anastomosis of the IAA ($n = 1$) or patch augmentation of a direct anastomosis ($n = 1$). A third patient with Taussig–Bing anomaly expired secondary to hemorrhage and low cardiac output following direct anastomosis of IAA, arterial switch with closure of ASD and VSD. All these early deaths occurred following one-stage repair.

There have been 10 late deaths (7 in group I and 3 in group II), with 4 late deaths occurring before the second stage procedure. The etiology was low cardiac output ($n = 4$); sudden unexplained death ($n = 2$); pneumonia ($n = 1$); respiratory failure ($n = 1$); bleeding ($n = 1$); and rejection following orthotopic heart transplantation ($n = 1$).

The overall actuarial survival including early mortality was 92% at 1 year, 81% at 5 years, and 76% at 10 and 15 years (Fig. 4A). Actuarial survival curves for patients with staged or one-stage IAA repair are shown in Fig. 4B. There was 78%

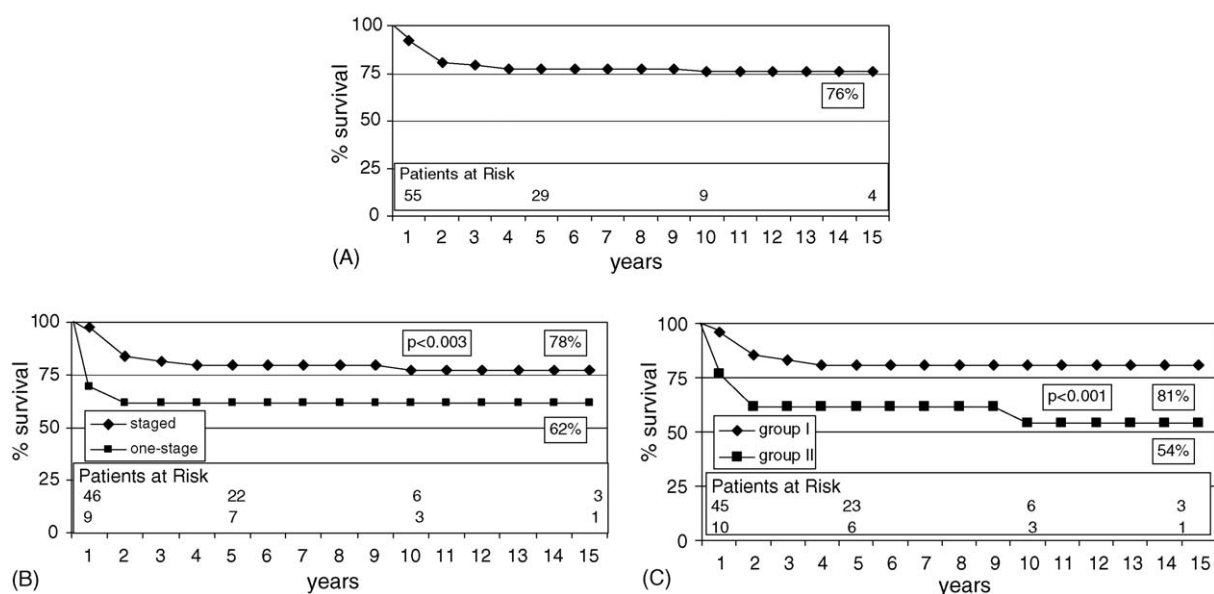


Fig. 4. Actuarial survival curves for patients with interrupted aortic arch repair: (A) overall; (B) in patients with staged and one-stage repair; and (C) in patients of group I and group II.

15-year survival probability for patients with staged IAA repair compared with 62% for patients with one-stage IAA repair ($p < 0.003$). Actuarial survival curves for patients in group I and group II are shown in Fig. 4C. There was 81% 15-year survival probability for children in group I compared with 54% for children in group II ($p < 0.001$). Risk factors for increased mortality by univariate analysis were as follows: (1) primary aortic anastomosis ($p = 0.03$), (2) presence of complex anomalies ($p = 0.05$), and (3) initial IAA repair performed before 1994 ($p = 0.05$). There were no risk factors identified by multivariate analysis for increased mortality.

3.2. Reoperation

Twenty patients underwent 27 reoperations (36%, 20/55; group I, 17 patients; group II, 3 patients) (Table 2). There was no death at reoperation. The time of reoperation ranged from 1 week to 9 years (mean, 29.1 ± 31.3 months).

Fifteen patients developed recurrent arch obstruction (group I, 14 patients; group II, 1 patient; $p < 0.002$). All had maximum pressure gradients more than 40 mmHg (mean, 51 ± 13 mmHg; range, 42–82 mmHg) at the site of the previous IAA repair. Two patients had successful balloon angioplasty while the remaining 13 patients had aortic arch augmentation with PTFE patch. Twelve patients (29%; 12/41) initially had arch reconstruction with the left carotid artery while three had a direct anastomosis (18%; 3/17; $p = 0.12$). Actuarial freedom from recurrent arch obstruction requiring reintervention was 74% at 15 years.

Four patients have undergone reoperation (7%; 4/55; group I) directly to relieve left ventricular outflow tract obstruction: subaortic fibromuscular and/or fibrous membrane resection, patch aortoplasty for relief supravalvar aortic stenosis. Patients who required reintervention ($n = 4$) underwent pre-operative echocardiographic examination and the peak LVOTO gradient was 73.8 ± 10.5 mmHg (range, 55–90 mmHg). At latest follow-up, the peak gradient in

Table 2
Reoperation

Cause	Group I	Group II
Recurrent coarctation	14	1
Resection of subaortic stenosis	4	0
Residual VSD	1	1
Heart transplantation	1	1
Reconstruction of RVOT with PTFE monocusp	0	2
RV–PA conduit	0	1
DeVega TV annuloplasty	0	1

PA: pulmonary artery; PTFE: polytetrafluoroethylene; RV: right ventricle; RVOT: right ventricular outflow tract; TV: tricuspid valve; VSD: ventricular septal defect.

these patients was 21.4 ± 8.1 mmHg (range, 12–36 mmHg). All of these patients have mild or no aortic insufficiency. Actuarial freedom from LVOT obstruction requiring reoperation was 92% at 15 years.

A residual VSD required closure in two patients. Diaphragmatic plication was performed in three patients because of left phrenic nerve palsy. Three patients from group II underwent reconstruction of right ventricular outflow tract with PTFE monocusp ($n = 2$) or Contegra conduit (Medtronic Inc., Minneapolis, MN, USA; $n = 1$) because of previous conduit obstruction ($n = 2$) or neo pulmonary artery stenosis ($n = 1$). One of these children also required DeVega tricuspid annuloplasty. Two patients developed end-stage cardiomyopathy and required heart transplantation (one in each group), of which one patient died (group II) 1 year later due to chronic rejection.

Actuarial freedom from any type of reoperation or intervention was 86% at 1 year, 69% at 5 years, and 60% at 10 and 15 years following the initial surgery (Fig. 5A). Actuarial freedom from any type of reoperation curves for patients with staged and one-stage IAA repair are shown in Fig. 5B. There was 49% 15-year freedom from reoperation probability for patients with staged IAA repair at presenta-

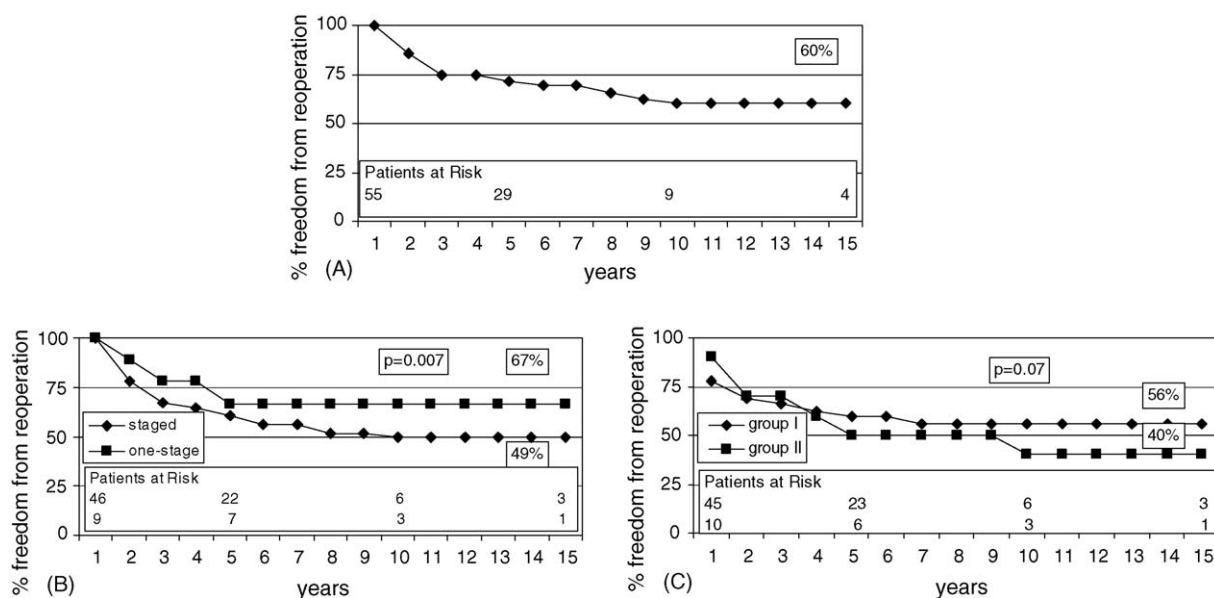


Fig. 5. Actuarial freedom from any type of reoperation curves for patients with interrupted aortic arch repair: (A) overall; (B) in patients with staged and one-stage repair; and (C) in patients in group I and group II.

Table 3
Results of surgical treatment in children with interrupted aortic arch

Author	Year	Number of patients	Method of repair	Method of AA repair	Mortality (overall) (%)	Freedom from reoperation
Irwin et al. [18]	1991	20	Staged (all)	PTFE tube graft (all)	25	0%
Karl et al. [3]	1992	55	One-stage (all)	Direct anastomosis (all)	24	69% at 4 years
Serraf et al. [5]	1996	79	One-stage (64) Staged (15)	Direct anastomosis (59) Direct anastomosis with PAu (8) Conduit (12)	30	No information
Mainwaring and Lamberti [20]	1997	27	Staged (all)	PTFE tube graft (all)	19	35% at 10 years
Atlaskal et al. [9]	1998	40	One-stage (19) Staged (21)	Direct anastomosis (24) Tube graft (10) Others (6)	50	No information
Fulton [16]	1999	72	One-stage (all)	Direct anastomosis (66) Others (6)	13	47% at 12 years
Schreiber et al. [19]	2000	94	One-stage (75) Staged (19)	Direct anastomosis (84) PAu (9)	38	40% at 15 years
Brown (present study)	2004	65	Staged (51) One-stage (14)	Left carotid swing down (43) Direct anastomosis (18) PTFE tube graft (2) PAu (2)	24 31 (before 1994) 19 (after 1994)	60% at 15 years

AA: aortic arch; PAu: patch augmentation; PTFE: polytetrafluoroethylene.

tion compared with 67% for patients with one-stage IAA repair ($p = 0.007$). Actuarial freedom from any type of reoperation curves for patients in groups I and II are shown in Fig. 5C. Univariate and multivariate analyses identified no variables as risk factors for reoperation.

3.3. Long-term follow-up results

The mean follow-up period was 5.5 ± 3.6 years, ranging 3 months to 20 years. Postoperative follow-up was available in 55 of the 57 survivors (97%). At last follow-up, none of the surviving children had cardiac symptoms and all reported a normal functional status. The majority (86%) of patients are in New York Heart Association (NYHA) class I, and 14% remain in class II. During the postoperative course there were no neurologic deficits, seizures, and growth disturbances in any patients. No restenosis or dilation was documented at the site of the arch reconstruction or at the level of the native aorta. Recent echocardiography in all survivors showed no evidence of dilation or obstruction at any level of the aorta and the peak gradient across the left ventricular outflow tract was 26.8 ± 10.5 mmHg (range, 10–40 mmHg).

4. Discussion

Successful surgical repair of IAA was first accomplished by Samson in 1955 in a patient with short-segment type A IAA [12]. A direct anastomosis was possible. The associated VSD was not closed at the time of the arch repair. The first successful one-stage correction of type A IAA was performed by Barratt-Boyes et al. [13] in 1970 where arch continuity was established using a synthetic conduit. In 1975, Trusler and Izukawa [14] reported correction of a type B IAA with a direct anastomosis of the ascending and descending aorta and closure of the VSD employing sternotomy with cardiopulmonary bypass in a 13-day-old baby.

Early experience with one-stage complete repair resulted in mortality of up to 65%, particularly in the presence of LVOTO [15]. Fortunately, mortality for patients having primary complete repair of IAA has decreased in parallel with improvements in results of other primary neonatal reconstructions for complex cardiac lesions over the past 15 years [3,9,16]. From a highly lethal condition where success was documented by individual cases [17], to small series [2,18] in which 50% of the infants survived, to more recent reports [5,9,19–21] with more improved outcomes as the surgical treatment continues to improve. Neonatal operative experience, early intubation, and prostaglandin infusion have helped to lower the operative mortality [4,19]. The clinical focus has now moved from early operative survival to medium- and long-term outcomes [5,9,19] (Table 3).

Despite these improvements, substantial operative mortality persists, and a number of late complications have been reported [18,5,9]. Complete primary repair has obvious appeal, but the complexities encountered in achieving this goal could be associated with a higher mortality. Because of our early interest in staged repair, we further evaluated the current role of this strategy when compared to primary total correction. Most of the problems associated with one-stage and primary complete repair have been defined in previous reports [18,3,5,9,19,20]. As experience with these procedures has grown, problems have decreased while others have persisted or even increased with longer follow-up.

Currently the one-stage approach is the preferred therapy for IAA and VSD at most institutions. There are advantages to this approach over the staged approach which includes fewer reoperations, avoidance of pulmonary artery banding which could accelerate subaortic stenosis and the decreased need for future arch reconstruction. Excellent results have been reported in a number of single-institution series [22]. The Congenital Heart Surgeons Society's multi-institutional study reported a 35% operative

mortality rate [4], suggesting that one-stage strategy does not always yield the most optimal outcome [5].

Associated complex anomalies, such as truncus arteriosus, double outlet right ventricle, carried a high risk [9,23]. Our own 46% overall mortality rate for repair of IAA with complex anomalies (group II) tends to support those findings.

Techniques for repair of the aortic arch have also been debated. Direct anastomosis of the ascending to the descending aorta with homograft or autologous pericardial patch augmentation has been advocated by several authors [4,5]. Others [3,6,20] favored the direct anastomosis without patch augmentation. Sell et al. [6] indicated direct anastomosis and earlier date of surgery were incremental risk factors for recurrent or persistent aortic arch stenosis. Our experience with staged repair of IAA indicates that using the left carotid artery as an autologous conduit for aortic arch continuity can be safely applied in these patients even in the presence of other intracardiac anomalies. Sacrifice of the left carotid artery for arch reconstruction does not appear to result in any adverse neurologic sequelae or growth disturbances during the follow-up period.

Bronchial compression is an unusual but consistent and troublesome complication observed after direct anastomosis or prosthetic graft interposition [19]. If the descending aorta is anastomosed more proximally on the ascending aorta, excessive tension between the two aortic components can lead to bronchial compression. This complication could potentially be avoided with anterior patch augmentation of the anastomosis. In our series, no patient has developed left bronchial compression.

The left ventricular outflow tract has been recognized as an important predictor of non-survival and reoperation in patients with IAA [5,6,16]. Mainwaring and Lamberti [20] described reoperations to relieve LVOTO in 18% of their survivors. Patients with IAA have posterior deviation of the conal septum, which morphologically can cause subaortic stenosis and even aortic annular hypoplasia leading to varying degrees of left ventricular outflow tract obstruction.

There is no general agreement as far as the need for primary intervention on a narrow LVOTO is concerned. Jonas et al. [4] does not recommend any intervention unless the subaortic stenosis is extreme. A relatively simple method of prevention of the LVOTO development is based on placement of the patch for VSD closure to the left side of the conal septum without myectomy. This technique was also recommended by Luciani et al. [22].

Balloon angioplasty for recurrent aortic arch stenosis after surgical repair has become the method of choice during the past decade. Although long-term data are still unavailable and the procedure is clearly not free of complications. In our experience, the recurrence of stenosis at the site of anastomosis which is extremely close to the origin of the left subclavian, making the patch angioplasty challenging and/or stenting impractical. Thus, balloon angioplasty for recurrence after primary anastomosis in the neonate can be challenging, as is surgical patch angioplasty. Roussin et al. [24] described that among the eight patients who required balloon angioplasty for recurrence, two required an early second angioplasty procedure and two have significant residual gradients in early follow-up. Late patch enlargement of the left carotid swing down anastomosis to the descending

aorta has been very successful in our hands and has had low morbidity and low reoccurrence rates.

In summary, our experience with staged repair of IAA for the past 20 years indicates that using left carotid artery swing down can be safely applied in IAA patients with and without other intracardiac anomalies. Use of the left carotid for arch reconstruction reduce early mortality and avoids several of the problems encountered with one-stage repair like difficult recurrent arch obstruction and left bronchial compression. Pulmonary banding used in staged repair has not increased the development of subaortic stenosis and may decrease subaortic stenosis. Our experience during the last 10 years shows a reduction in mortality from 31% to 19%. Associated anomalies play an important role in the outcomes. The long-term probability for reoperation or reintervention remains high regardless of the operative technique chosen.

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Editorial comment

Interrupted aortic arch

Interrupted aortic arch is a rare condition accounting for about 1% of infants with critical congenital heart disease. The arch interruption may be distal to the left subclavian artery (Type A), between the left carotid and left subclavian arteries (Type B) or between the innominate and left carotid arteries (Type C) [1]. Type B is the commonest and Type C is very rare. Associated defects such as a Ventricular septal defect (VSD) and Persistent ductus arteriosus (PDA) are almost always present and these constitute a non-complex group. Other associated lesions such as truncus arteriosus, transposition of the great vessels, Taussig-Bing double right ventricle and univentricular heart constitute a complex group. Left ventricular outflow tract obstruction may be present initially or develop later. The aortic valve is bicuspid in about 30% of cases and an aberrant right subclavian artery originating from the descending aorta is quite common, particularly in Type B.

Early in life the duct shuts and this results in severe congestive failure and metabolic acidosis. The great majority of patients present within the first four weeks of life. Before the availability of Prostaglandin, urgent operation was necessary; however, with Prostaglandin the baby can usually be stabilised before surgery. Pre-operative assessment with echocardiography confirms the diagnosis and establishes the presence of associated defects. It is 30 years since Trusler and Izukawa [2] showed how to correct the simple type through the midline with direct anastomosis of the descending to ascending aorta and closure of the VSD. An increasing number of surgeons have used this approach with a lateral thoracotomy approach being reserved for some of the complex group such as univentricular heart.

In that interrupted aortic arch can be considered to be an exaggerated type of coarctation, there is some illogicality in that the traditional approach for repair of coarctation and VSD has been through a left lateral thoracotomy together with pulmonary artery banding. In recent years, learning from the experience with interrupted arch repair, more surgeons are starting to treat coarctation and VSD through the midline. It therefore comes as somewhat of a surprise to see a large series of neonates with interrupted aortic arch being treated with a two-stage approach as reported in this journal [3]. They report 52 out of 65 neonates having a staged repair with the initial arch repair and pulmonary banding through the left chest. Their early mortality in the non-complex group is low although there is some cross-over between groups and it is not clear why they used a one-stage

repair in 14% of the non-complex group. It is clearly important to end up with live patients, but such an experienced surgeon would probably have had good results with a one-stage procedure in the non-complex group.

It is interesting to see their extensive use of the left carotid turn-down technique which helps to avoid tension. The left carotid artery in a 3.0-kg neonate with Type B interrupted aortic arch is usually about 4.0 mm in diameter and this is adequate for a new aortic arch. Brown et al. have shown that this can be used safely although there is a 33% incidence of late stenosis at the anastomotic site. It has been shown previously that a new arch formed by the left carotid artery grows to about 80% of normal size, which is quite adequate and without a significant gradient across it [4]. Whichever method of repair is used, it is very important to remove all duct tissue as failure to do so predisposes to subsequent aortic narrowing. The use of synthetic tube grafts such as Gore-Tex should be avoided as obviously they cannot grow and further surgery is inevitable.

Although the early mortality in the non-complex group of patients undergoing a two-stage approach is low in Brown's series, there is a disconcerting subsequent outcome, as four patients needed their second stage within two weeks and four died before the second procedure. Furthermore there was a significantly lower (49%) 15-year freedom from re-operation with staged repair than with a one-stage approach (67%).

Brown et al. suggest that direct anastomosis is a risk factor for increased mortality. However, this is not a fair conclusion as the patients having direct anastomoses who died were complex (two with truncus arteriosus and one Taussig-Bing). Obviously the mortality will be higher in a complex group of patients than in non-complex, most of whom had staged repair.

It is encouraging that the results of surgery for interrupted aortic arch are steadily improving with an early mortality in the region of 10% [5,6]. This improvement relates to many other factors such as improved pre-operative state, anaesthesia, bypass techniques and post-operative care. In particular with modern small aortic cannulae, the ability to continue some perfusion through the innominate artery during repair of the aortic arch rather than having to use deep hypothermia and circulatory arrest is a major advance.

Most surgeons would agree that for the non-complex group, a single stage procedure through the midline which